Ruptured Congenital Sinus of Valsalva Aneurysm

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A previously healthy 39-year-old male presented with a 2-week history of progressive exertional dyspnea with fatigue. On examination, he had a grade 4/6 continuous cardiac murmur, loudest however, in systole. Transthoracic echocardiography revealed a ruptured right sinus of Valsalva aneurysm (SVA) into the right atrium (RA) (Fig. 1). The patient subsequently underwent surgical patch repair and was discharged home on the fourth postoperative day.

Dilatation of all three sinuses of Valsalva may be noted with aging, and probably with hypertension.[1,2] Other etiologies of enlargement of all three sinuses include Marfan's syndrome, annuloaortic ectasia, and other connective tissue diseases,[3–5] syphilis,[6] and ankylosing spondylitis.[6,7]

Aneurysm of one or two of the three aortic sinuses of Valsalva is an unusual occurrence. The most common etiology is from spread of infective endocarditis and formation of a ring abscess.[8,9] A noninfective etiology is very uncommon, and is felt to be due to a congenital absence of media in the wall of the aorta within the sinus of Valsalva.

Congenital SVA occurs mostly in males (4:1 male:female ratio), with presentation typically in young adults.[10,11] The aortic root media is in discontinuity with the aortic valve annulus,[12,13] thus the aneurysm orifice is in proximity to the floor of the sinus of Valsalva. In addition, a congenital SVA will have an aneurysm channel (finger-like projection), generally not found in those with an infective etiology. The right coronary sinus is involved most often (69% incidence), followed by the noncoronary sinus (26% incidence), and rarely the left coronary cusp (5% incidence).[14] Congenital involvement of more than one sinus may rarely occur.[15] A supracristal ventricular septal defect (VSD) is often associated with a right SVA.

Expansion of an SVA will follow the path of least resistance; therefore, it will usually encroach upon the right heart. If the SVA then ruptures, a left-to-right shunt will form. A right SVA will rupture into the right ventricle or the RA, as occurred with this patient. A noncoronary SVA will almost always rupture into the RA.[16] Rarely, a congenital SVA will dissect into the interventricular septum, where it may then later rupture into the right ventricle (RV) or left ventricle (LV).[17–19] Rare rupture into the pericardial space is associated with sudden death.[20]

Reported cases of ruptured SVA are more numerous than those that are unruptured. Most unruptured SVAs probably produce no symptoms, although rarely may cause RV outflow tract obstruction,[21] heart block,[22] or LV outflow tract obstruction.[23]

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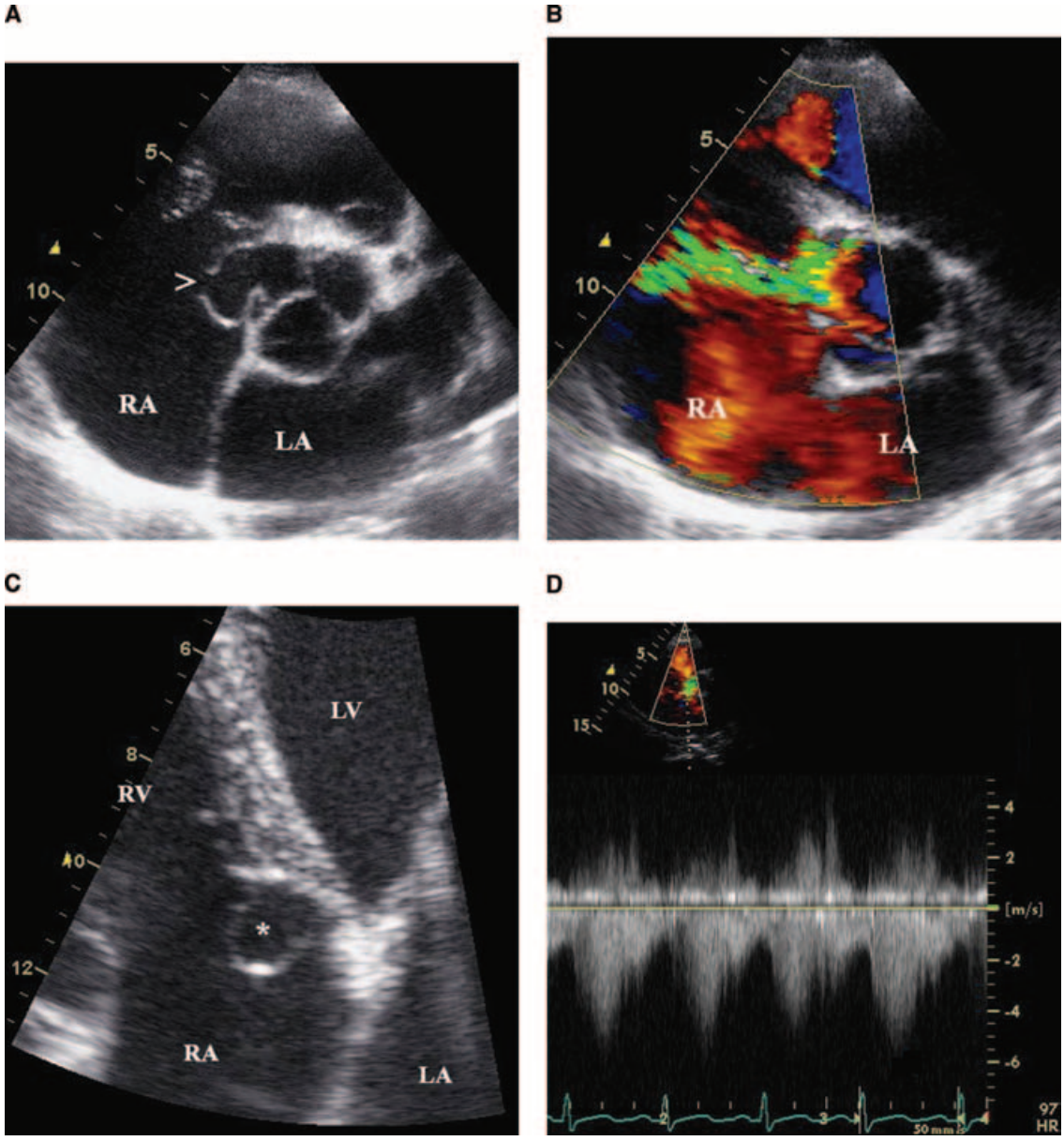

Figure 1. *Transthoracic echocardiographic imaging of a ruptured sinus of Valsalva aneurysm.* **A.** *Parasternal short-axis at a level just above the aortic valve reveals the site of rupture within the right coronary sinus (>).* **B.** *Color flow Doppler at the same level demonstrates turbulent yellow-green flow from the ruptured sinus of Valsalva into the right atrium (RA). This turbulence was noted throughout the entire cardiac cycle.* **C.** *Close-up imaging of an apical four-chamber image reveals the aneurysm (∗) within the RA above the tricuspid septal leaflet.* **D.** *Continuous wave Doppler from a parasternal short-axis view (same level as in Fig. 1A and 1B). Flow is noted throughout the cardiac cycle, but is highest during systole. LA = left atrium; LV = left ventricle; RV = right ventricle.*

A new continuous murmur in a previously healthy patient is classical presentation for ruptured SVA.[24] The murmur will be generally louder either in systole (as was in this case) or in diastole. In distinction from this, the continuous murmur of patent ductus arteriosus will peak about the second heart sound.[25,26] The continuous murmur of a coronary artery fistula may be maximal at an unusual chest position.[27] Patent ductus arteriosus and coronary artery fistula, however, are readily distinguished from ruptured SVA by echocardiography.

The continuous murmur of ruptured SVA is distinctly different from that of the murmur of a VSD. A small perimembranous VSD may have only an early systolic or holosystolic murmur. The murmur will end before the onset of diastole.[28]

An echocardiogram is often performed to evaluate a continuous murmur. An SVA (ruptured or unruptured) is well visualized from a parasternal short-axis view at the level of the aortic root. An unruptured SVA most often has a thinner wall and is larger than the other sinuses. If ruptured into the right heart, color Doppler will demonstrate a continuous turbulent jet within the aneurysm flowing into the receiving chamber. Also, tricuspid valve fluttering and diastolic pulmonic valve opening may be noted.[29] As increased blood flow enters the pulmonary artery, left atrium (LA), and LV, LV volume overload will be seen with significantly sized shunts. In addition to LV volume overload, rupture into the RA causes volume overload of the RA and RV also, and rupture into the RV causes volume overload of the RV. Because volume overload of both ventricles occurs when ruptured into the right heart, paradoxical ventricular septal motion usually is not found.

If an SVA ruptures into the LV, a jet occurring only in diastole will be noted. This jet may initially appear to be aortic regurgitation. In addition, this diastolic jet is to be differentiated from aortico-left ventricular tunnel, in which the tunnel opens to the aorta above the coronary ostium, and the ruptured SVA opens to the aorta below the coronary ostium. Mild aortic regurgitation may be seen associated with SVA ruptured into the right heart, but severe aortic regurgitation should alert one to look for rupture into the LV or bacterial endocarditis affecting the aortic valve.

An SVA is to be differentiated from the more common aneurysm of a membranous VSD. Close short-axis imaging of the LV outflow tract and aortic root helps identify the VSD aneurysm originating below the aortic annulus, and the SVA above the aortic annulus. If a left-to-right shunt is present (ruptured SVA vs perimembranous VSD), differentiation is determined by timing shunt flow to the electrocardiogram. Systolic flow occurs only with a VSD, and both systolic and diastolic flow with a ruptured SVA.

In addition to a perimembranous VSD, an SVA is to be differentiated from a coronary arteriovenous (AV) fistula. Importantly, parasternal short-axis imaging will help identify the origin of normal-sized coronary arteries in SVA. A dilated coronary artery, notably seen at its origin from the aorta, is suggestive of a coronary AV fistula.

References

1. Silver MA, Roberts WC: Detailed anatomy of the normally functioning aortic valve in hearts of normal and increased weight. *Am J Cardiol* 1985;55:454–461.
2. Roman MJ, Devereux RB, Kramer-Fox R, et al: Two-dimensional echocardiographic aortic root dimensions in normal children and adults. *Am J Cardiol* 1989;64:507–512.
3. Roberts WC, Honig HS: The spectrum of cardiovascular disease in the Marfan syndrome: A clinicomorphologic study of 18 necropsy patients and comparison to 151 previously reported necropsy patients. *Am Heart J* 1982;104:115–135.
4. Waller BF, Reis RL, McIntosh CL, et al: Marfan cardiovascular disease without the Marfan syndrome. Fusiform ascending aortic aneurysm with aortic and mitral valve regurgitation. *Chest* 1980;77:533–540.
5. Goldstein SA, Lindsay J: Thoracic aortic aneurysms: Role of echocardiography. *Echocardiography* 1996;13(2):213–232.
6. Schlant RC, Gonzalez EB, Roberts WC: The connective tissue diseases. In: Alexander RW, Schlant RC, Fuster V (eds): *Hursts's The Heart*. New York, McGraw-Hill, 1998, pp. 2284–2285.
7. Bulkley BH, Roberts WC: Ankylosing spondylitis and aortic regurgitation. Description of the characteristic cardiovascular lesion from study of eight necropsy patients. *Circulation* 1973;48:1014–1027.
8. Shaffer EM, Snider AR, Rosenthal A: Sinus of Valsalva aneurysm complicating bacterial endocarditis in an infant: Diagnosis with two-dimensional and Doppler echocardiography. *J Am Coll Cardiol* 1987;9:588–591.
9. Arnett EN, Roberts WC: Valve ring abscess in active infective endocarditis. Frequency, location, and clues to clinical diagnosis from the study of 95 necropsy patients. *Circulation* 1976;54:140–145.
10. Aletras H, Bjork VO, Cullhed I, et al: Ruptured congenital aneurysm of the sinus of Valsalva with ventricular septal defect. *Thorax* 1963;18:127–135.
11. Perloff JK: Sinus of Valsalva—Right heart communications due to congenital aortic sinus defects. *Am Heart J* 1960;59:318–321.
12. Edwards JE, Burchell HB: The pathologic anatomy of deficiencies between the aortic root and the heart including aortic sinus aneurysms. *Thorax* 1957;12:125–139.
13. Edwards JE, Burchell HB: Specimen exhibiting the essential lesion in aneurysm of the aortic sinus. *Proc Staff Meet Mayo Clin* 1956;31:407–412.
14. Adams JE, Sawyers JL, Scott HW: Surgical treatment for aneurysms of the aortic sinuses with aorticoatrial fistula. *Surgery* 1957;41:26–42.
15. Chamsi-Pasha H, Musgrove C, Morton R: Echocardiographic diagnosis of multiple congenital aneurysms of the sinus of Valsalva. *Br Heart J* 1988;59:724–726.
16. Sakakibara S, Konno S: Congenital aneurysms of the sinus of Valsalva—Anatomy and classification. *Am Heart J* 1962;63:405–412.

17. Chen WW, Tai YT: Dissection of interventricular septum by aneurysm of sinus of Valsalva. A rare complication diagnosed by echocardiography. *Br Heart J* 1983;50:293–295.
18. Engel PJ, Held JS, Bel-Kahn JV, et al: Echocardiographic diagnosis of congenital sinus of Valsalva aneurysm with dissection of the interventricular septum. *Circulation* 1981;63:705–711.
19. Guler N, Eryonucu B, Tuncer M, et al: Aneurysm of sinus of Valsalva dissecting into interventricular septum: A late complication of aortic valve replacement. *Echocardiography* 2004;21(7):645–648.
20. Roberts WC: Congenital cardiovascular abnormalities usually "silent" until adulthood: Morphologic features of the floppy mitral valve, valvular aortic stenosis, discrete subvalvular aortic stenosis, hypertrophic cardiomyopathy, sinus of Valsalva aneurysm, and the Marfan syndrome. *Cardiovasc Clin* 1979;10(1):442–453.
21. Kerber RE, Ridges JD, Kriss JP, et al: Unruptured aneurysm of the sinus of Valsalva producing right ventricular outflow obstruction. *Am J Med* 1972;53;775–783.
22. Segab C, Davy JM, Scheuble C, et al: Atrioventricular block disclosing an isolated congenital aneurysm of the sinus of Valsalva, extending into the septum and not ruptured. *Arch Mal Coeur* 1981;74:1233–1239.
23. Rothbaum DA, Dillon JC, Chang S, et al: Echocardiographic manifestations of right sinus of Valsalva aneurysm. *Circulation* 1974;49:768–771.
24. Sakakibara S, Konno S: Congenital aneurysm of the sinus of Valsalva: A clinical study. *Am Heart J* 1962;63:708–719.
25. Kieffer SA, Winchell P: Congenital aneurysms of the aortic sinuses with cardioaortic fistula. *Dis Chest* 1960;38:79–96.
26. Neill C, Mounsey P: Auscultation of patent ductus arteriosus with a description of two fistulae simulating patent ductus. *Br Heart J* 1958;20:61–75.
27. Sakakibara S, Yokoyama M, Takao A, et al: Coronary arteriovenous fistula; nine operated cases. *Am Heart J* 1966;72:307–314.
28. Feruglio GA, Gunston RW: Intracardiac phonocardiography in ventricular septal defect. *Circulation* 1960;21:49–58.
29. Weyman AE, Dillon JC, Feigenbaum H, et al: Premature pulmonic valve opening following sinus of Valsalva aneurysm rupture into the right atrium. *Circulation* 1975;51:556–560.